

Total Nasal Skeletal Reconstruction Disfigured by Granulomatosis with Polyangitis (Wegener Granulomatosis)

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Background: Nasal deformity is a common disorder in Wegener granulomatosis, which is produced by polyangitis of medium-sized vessels. This process may cause necrosis and destruction of the nasal mucosa and osteocartilaginous framework. At this time, there is a lack of published data on nasal deformities associated with Wegener granulomatosis.

Methods: We present a 53-year-old woman with completely necrotized nasal lining and osteocartilage. The patient had a history of granulomatosis with polyangitis that was currently in remission. We reconstructed the patient's nose with fifth and sixth rib cartilages over 3 successive surgeries over a 2-year period.

Results: During the 3-year follow-up period, neither infection nor significant cartilage graft resorption was observed, and the aesthetic result was acceptable both to the patient and her physicians.

Conclusions: In spite of contamination to the nasal lining and immune-compromising medications, nasal reconstruction is usually successful when considering these factors. Access to strong and abundant fifth or sixth rib cartilage presents an opportunity to reconstruct nasal destruction due to iatrogenic or autoimmune processes. (*Plast Reconstr Surg Glob Open* 2015;3:e308; doi: 10.1097/GOX.0000000000000278; Published online 19 February 2015.)

Described in 1936 by German pathologist Friedrich Wegener, granulomatosis with polyangitis is the most common upper airway granulomatosis disease. At this time, there

is a paucity of literature concerning nasal reconstruction in patients with Wegener granulomatosis (WG).¹⁻³ Surgeons are reluctant to perform nasal reconstruction in these patients for several reasons, including fear of local and systemic flare up, fear of contaminated nasal mucosa, the possibility of redestruction and infection of grafted structures, and use of prolonged anti-inflammatory medications.¹ Severe nasal disfigurement may have serious psychological consequences on a victim's behavior. A high rate of remission and disease control of WG with modern medical therapy has opened the door for reconstructive intervention into this condition. Here, we present the results of a woman with a history of WG and completely necrotized nasal lining and osteocartilage who underwent successful nasal reconstruction with rib cartilage.

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CASE REPORT

A 53-year-old woman was presented to our practice for total nasal reconstruction. Sixteen years ago, at age 37, she was referred to otolaryngology with a complaint of nasal obstruction. At that time, she underwent inferior turbinectomy and septoplasty for correction of the obstruction. Her symptoms returned 5 years later, with nasal mucosa necrosis, extensive septal perforation, dorsal collapse, and hoarseness. Autoimmune disease was suspected, and a biopsy was performed, which revealed necrotizing granulomatous inflammation. All laboratory tests were within normal limits. Cyclophosphamide and trimethoprim/sulfamethoxazole were initiated at this time, which was later combined with an oral corticosteroid. Eight years ago, after complete remission (at age 45), an attempt for nasal reconstruction with an iliac bone graft was conducted. The bone graft was resorbed gradually with severe destruction and collapse of the nasal skeleton. Despite care from a team consisting of a psychiatrist, a rheumatologist, and an otolaryngologist, the patient became severely depressed and had several suicide attempts and was eventually referred to our practice for total nasal reconstruction.

On physical examination, we observed extensive perforation of the septum, complete lack of bony and cartilaginous framework in the nose, and contraction of covering skin and mucosal lining (Fig. 1). For the first operation, we harvested cartilage from the fifth rib before addressing the nose. Cartilage was removed through a 2-cm incision on the right inframammary line, 2 cm from midline. A total of 4 cm of harvested rib cartilage was divided into several 2-mm-thick strips and immersed in saline solution before beginning the nasal opera-

tion. We used an open procedural approach with caution toward avoiding the inside of the nasal cavity to prevent contamination. Augmentation of the dorsum was achieved with a strong dorsal cartilage graft and columella strut sutured together to make an L-shaped scaffold, 2 lateral wall grafts, and 2 alar strut grafts.

We conducted wide skin undermining through a step columellar incision. Two layers of 4.0×0.5 cm cartilage strips were used on the dorsum and one 2.0×0.4 cm strip was used for each alar strut. A piece of 1.8×0.4 cm cartilage was inserted as a columella strut and 2-layer cap graft.

After 1 year of follow-up, the patient was on medication and experienced no recurrence of nasal deformity. Grafted cartilages remained in place with no sign of resorption. Although she could easily breathe through her nose and with the aesthetic improvement was in a better psychological condition, the nose was short with a high tip and had a shortage of the lateral wall. At this time, a second operation was conducted via a left 2-cm submammary line incision, and 4 cm of cartilage from the sixth rib was harvested and again divided into 2-mm-thick strips. Two strips of $22 \times 6 \times 2$ mm cartilage were inserted into the lateral walls, and 2 strips of $15 \times 4 \times 1$ mm cartilage were inserted at the alar area. Six months later, a third procedure—nasal skin undermining and release of scar tissue—was performed to further correct the foreshortened nose and adjust the remaining alar rim retraction. An extended tip graft and 2 alar rim grafts were applied using the banked cartilage of the previous operation. The lateral wall grafts were trimmed conservatively to decrease the width of the nose. After 3 years of follow-up from the third operation, the

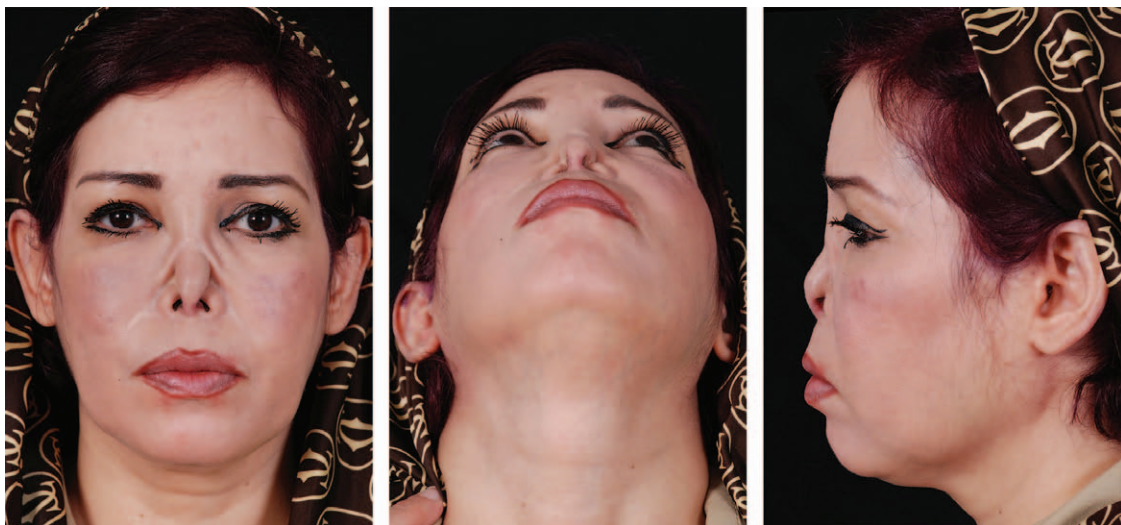


Fig. 1. Patient referred to our clinic at age 47 with total nasal skeletal destruction.



Fig. 2. Patient has normal breathing and aesthetically acceptable appearance 3 years after last operation.

patient remains in remission and can breathe normally through her nose. The reconstructed nose is stable and aesthetically acceptable (Fig. 2).

CONCLUSIONS

We believe that rib cartilage should be the gold standard for reconstruction of this type of nasal deformity, including cases of trauma, resected secondary operations, congenital anomalies, and auto-immune-destructed nasal framework.⁴ The structure of rib cartilage is stronger than the other autologous grafts, with predictable warping.

Although immune suppressants may inhibit the normal wound healing process of WG, and the nature of WG itself includes contamination and scarred nasal lining, it seems that intervention with aggressive reconstructive modalities may help patients with WG and should be considered a strong option for surgical intervention when in remission and we should consider that this disease is a life-long process.

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PATIENT CONSENT

The patient provided written consent for the use of her image.

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